SYRINGOCYSTADENOMA PAPILLIFERUM

REPORT OF A CONGENITAL LESION OF ECCRINE ORIGIN

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Abstract — We are reporting on a patient with a large linear syringocystadenoma papiliferum present at birth over the anterior surface of the left leg. Histologic findings and the results of immunohistochemical staining with monoclonal antibodies CEA and IKH-4 suggest derivation from eccrine glands.
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INTRODUCTION

Syringocystadenoma papilliferum is a benign adnexal neoplasm which usually shows apocrine differentiation. The most common location is the head and neck region, often in the setting of an organoid nevus (nevus sebaceus of Jadassohn). Clinically the lesion is verrucous with a moist surface indicative of the secretory elements within the underlying tumor. Reports of syringocystadenoma on the extremities and independent of an organoid nevus are rare. The anatomical distribution of syringocystadenoma papilliferum in regions which do not normally contain apocrine glands led some pathologists to conclude that it is either an adenoma of eccrine glands in which the eccrine glands are no longer recognizable or it arises from the ducts of a gland intermediate between eccrine and apocrine (1). However, Pinkus believed that the syringocystadenoma papilliferum is a hamartoma which is derived from mature apocrine glands and in occasional cases, from eccrine glands (2). We report a case of syringocystadenoma papilliferum present since birth and arising on the anterior aspect of the leg. We also present evidence for an eccrine derivation of this neoplasm.

CASE REPORT

A 32 year old woman presented with a linear lesion of the left anterior leg present since birth. The lesion had recently chnaged, becoming larger, and measured 19 cm in length and 3.5 cm in width (Fig. 1) The surface was verrucous and moist. A portion of the lesion was removed by surgical excision for hisotlogic examination. The epidermis showed verrucous hyperplasia. Numerous dilated and branching ductal structures lined by two layers of cuboidal cells were present in the underlying dermis, opening into the overlying epidermis in multiple areas (Fig. 2). The tubular sturctures were surrounded by a fibrovascular stroma containing numerous plasma cells (Fig. 3). The deep dermis contained numerous eccrine sweat coils, half secretory and half ductal in nature. There were no identifiable apocrine glands (Fig. 4). Immunohistochemical staining for CEA utilizing paraffin embedded tissue showed a positive staining pattern in both the branching tubular structures and the underlying eccrine glands. Sections of the tumor were also stained for IKH-4, a monoclonal antibody initially

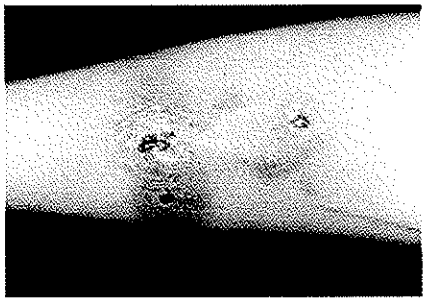

Fig. 1. Clinical photograph of the patient.

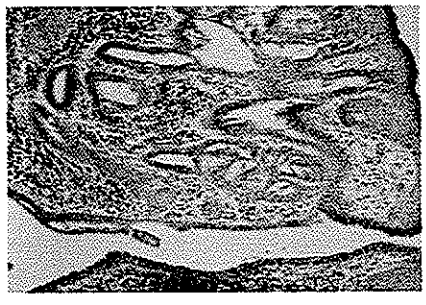

Fig. 2. Histological appearance of the lesion.

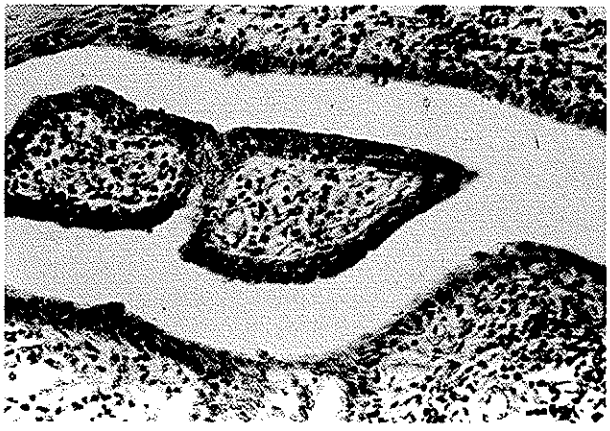

Fig. 3. Tubular structures surrounded by a fibrovascular stroma containing numerous plasma cells.

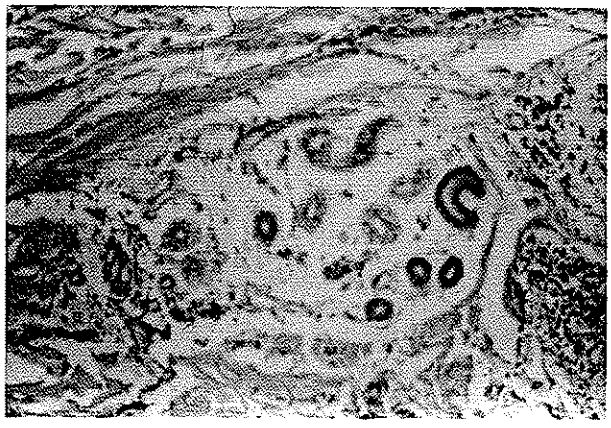

Fig. 4. Deep dermis containing numerous eccrine sweat coils.

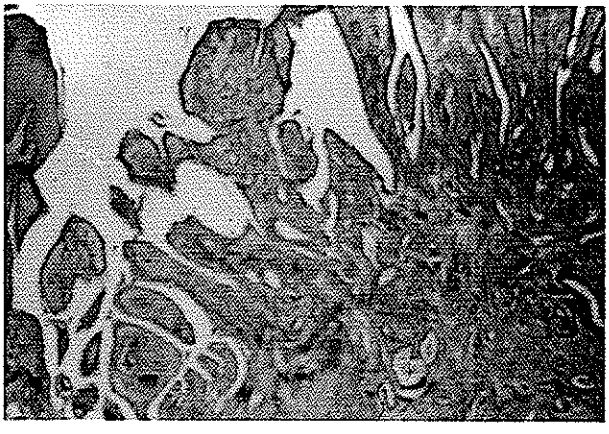

Fig. 5. Branching ductal structures of the syrimgocystadenoma papilliferum stained positvely IKH-4, as did the underlying eccrine glands.

produced for the identification of melanocytes, but also found to stain epithelial structures. It was found thatIKH-4 stains eccrine glands while apocrine glands are unractive. As a control, sections of axillary skin and organoid nevus was stained with IKH-4 (Fig. 5). The eccrine gland were positive while the apocrine glands present in the same sections were unreactive. Branching ductal structures of the syringocystadenoma papilliferum stained positively with IKH-4, as did the underlying eccrine glands.

DISCUSSION

While the majority of syringocystadenoma papilliferums develop within organoid nevi, some have been described arising de novo (3). The location of some lesions on the extremities where apocrine glands are not present suggest that a minority of syringocystadenoma papilliferums arise from eccrine glands. The absence of apocrine glands and abnormal development of the pilosebaceous structures in our case rules out the possibility of an underlying organoid nevus. The fact that our patient had a previous lesion present at birth and the numerous intact eccrine glands deep in the dermis raise the possibility of a pre-existing congenital eccrine nevus in the area. Some authors have previously proposed an eccrine origin for syringocystadenoma papiliferum on the basis of electron microscopic findings (4) others have found positive staining for gross cystic disease fluid protein, a marker of mammary glands and apocrine differentiation (5). Staining with monoclonal antibodies IKH-4 provides evidence for eccrine origin in this lesion. The IKH-4 stained eccrine but not apocrine glands in axillary skin and in an organoid nevus. These results, along with the absence of underlying apocrine glands, suggest that in this lesion of syringocystadenoma papilliferum is derived from eccrine glands.

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